SUMMARY. *Diagnosis.* Chronic and parenchymatous nephritis; acute general infection resulting in infarction of the lungs; double phlebitis; nephrolithiasis of both kidneys.

Prognosis. On two occasions during the course of the year the patient was manifestly close to death: the first time just before the operation for decapsulation of the kidneys, and the second time after the attack of kidney colic, when there was almost complete suppression of the urine.

It is manifestly impossible to be at all sure about the outcome in this case, but the whole course of the disease shows that there must have been, and must be still, a certain amount of healthy kidney tissue left. Therefore, despite the desperate illness through which the man has passed, and despite the fact that the excretion of albumin and casts has not changed, one may venture to hope that there are an indefinite number of years before this man, during which he may live and possibly still do good work. My experience, especially of late years, has taught me the great uncertainty and wide variations in the course of kidney cases. No general rule can be laid down, but a complete study of the individual equation will sometimes help us in establishing a prognosis that will prove approximately correct.

So far as the treatment in this case is concerned, drugs exerted practically no effect, and the same may be said as regards diet, with the exception that exclusion of salt from all articles of food unquestionably had a great influence upon the œdema.

The Edebohls operation in this case was of doubtful utility. My impression is that it did no good beyond relieving tension at the time.

Venesection acted wonderfully as a diuretic, but produced no permanent result.

A last point, and one which is too often forgotten, is that patients suffering from parenchymatous nephritis are embarrassed rather than benefited by subcutaneous injections of salt solution. In fact, we are to be on our guard about using this procedure, because at times it may prove exceedingly dangerous.

FATAL OBLITERATING ENDOPHLEBITIS OF THE HEPATIC VEINS.

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(From Prof. Chiari's Pathologico-anatomical Institute, Prague.)

A DISEASE characterized by a gradual proliferation of the lining wall of the hepatic veins and resulting obstruction, with all its disastrous consequences to the liver and portal system, was mentioned for the first time by Budd in 1846 in his treatise on the liver. Since that time numerous similar cases have appeared in the literature, some 23 in all, the majority of which have been published in the last ten years. The German, French, Swiss, Italian, and English literature contains examples of this obliterating endophlebitis of the hepatic veins, but as yet no case has appeared in American scientific or clinical publications. We can hardly believe that cases of this nature have not at some time presented themselves in the clinics of our country. Moreover, when we consider that the outcome of the disease is invariably fatal, the conviction is strengthened that postmortem examinations upon such cases have also been made without the true cause of the pathological lesions and of death being disclosed. This is very easily explained and understood when we realize that, clinically, the cases mask themselves under the every-day garb of a Laennec cirrhosis, and, therefore, are not apt to attract particular consideration; and that at the autopsy, unless the hepatic portion of the inferior vena cava is opened, and the ostia of the hepatic veins there examined, the clinical diagnosis of cirrhosis of the liver may readily be declared to have been correct. To bring out more forcibly the point that this condition is overlooked, not only in the United States but elsewhere, and is by no means as rare as the paucity of reported cases would lead one to believe, let me call attention to the fact that five of the 23 cases below mentioned are reported from one pathological institute, that of Chiari, of Prague, and that since his thorough exposition of the subject, in 1899, two further cases have been found by him at autopsy. It is therefore to bring this condition to the notice of the clinician and pathologist that I publish a full report of a case of this disease. For I feel convinced that we cannot hope to obtain a clearer insight into its cause, or a recognition of its clinical aspects, except by a consideration of many more cases than have been studied and collated up to the present time. In order to give the reader a comprehensive idea of the symptomatology and pathology of a disease which is as yet so little known, I have prepared a tabular synopsis of all the cases which I have been able to find, and shall precede the report of the case which suggested the article, by a general survey of all the data at hand. The facts are in each case taken directly from the original reports, the evident omission of important data being due to lack of information furnished by the author.

Case.	Age and sex.	Clinical data.	Autopsy.	Microscopic data.	Conclusions of the author.
Budd, 1846	52 years ♂	....	Hepatic veins thickened and containing a "pseudomembrane;" thrombosis of portal veins; adhesive perihepatitis, pericarditis, and pleuritis.	....	No cause assigned for thickening of veins; the adhesive serositis attributed to irregular mode of life, probably alcoholism being meant.
Cohn, 1860	....	....	Widespread phlebitis of hepatic veins; thrombosis of portal veins; ascites; enlarged spleen.		
Frerichs (1), 1861	38 years ♀	Abdominal pain, sacculated peritoneal exudate, slight icterus, œdema of lower half of body.	Ostia of hepatic veins closed by complete or partly fenestrated septa and surrounded by thick connective-tissue sheath; liver smaller by a third, firmly bound down, the capsule thickened, sending firm bands into liver tissue.	....	The valve-like closure of hepatic veins due to a primary peritonitis causing a secondary perihepatitis with connective-tissue bands entering the liver tissue and pressing on the venous walls.
Frerichs (2)	45 years ♀	Ascites, diarrhœa, icterus, enlarged spleen.	One hepatic vein completely closed by a light red mass, the size of a cherry; one ending blind, with wall greatly thickened; a third containing adherent thrombi; the vena cava containing round plaques, its walls flattened together by connective-tissue bands; loosely attached thrombi in portal vein; liver somewhat small, connective-tissue masses on under surface, extending into parenchyma.	....	Adhesive phlebitis of hepatic veins due to chronic peritonitis.
Rosenblatt, 1867	27 years ♀	Sticking abdominal pain, ascites, distention of abdominal veins.	Ostia of hepatic veins entirely absent but represented by slight elevations and depressions of wall of vena cava; left lobe of liver tightly bound down, bands extending from capsule into parenchyma; primary carcinoma of liver with carcinomatous peritonitis.	....	Hepatic veins obstructed as result of fetal hepatitis.
Gee, 1871	17 mos. ♀	Appeared to have abdominal pain; ascites.	Hepatic veins closed off from vena cava by thin membrane, showing merely shallow dimples at these sites, many filled with tough thrombi; liver somewhat small, tough, with perihepatitis; signs of compensatory circulation.	....	Primary cirrhosis of liver, extending to capsule, especially about hepatic veins.
Schüppel, 1880	16 years ♀	Serofibrinous pleurisy, involving the right pleura (the only symptom mentioned).	Several hepatic veins of right lobe completely closed by organized thrombi; liver rather large; no perihepatitis; spleen very large; no ascites; organized thrombi in longitudinal sinus and both transverse sinuses of dura mater.	....	Possibly alteration of coagulability of blood, causing tendency to thrombosis.

Hainski, 1884	22 years ♂	Ill for one year. Diagnosis of chronic peritonitis with pressure on vena cava and bile-ducts by new growth of fibrous tissue.	Organized thrombi in hepatic veins, no large vein emptying into vena cava; the walls of hepatic veins and vena cava thickened and surrounded by connective tissue which extended into liver; liver somewhat small, especially the right lobe; tough, congested, granular, and containing grayish-brown nodules; moderate perihepatitis; ascites, chronic enlargement of spleen, purulent peritonitis.	The nodules were found to be due to circumscribed hyperplasias, in the nature of regenerative growth of parenchyma.	Obliteration of the hepatic veins following thrombosis; the primary lesion was a chronic peritonitis which caused cirrhosis.
Lange, 1886	39 years ♂	Gradual illness nine mos. after pregnancy, ascites, abdominal discomfort, anasarca, large liver; ill five and a half months in all.	Hepatic veins in liver appearing as connective-tissue bands, many absolutely obliterated, some containing thrombi; one at a point of juncture of two veins held a calcareous nodule the size of a cherry; the inferior vena cava showing longitudinal thickening and numerous ostia of hepatic veins blocked; some branches of portal vein outside of liver thickened; liver normal size, congested, showed dark red and circumscribed yellowish areas; localized perihepatitis on upper surface posteriorly; ascites.	Hepatic veins showed thickening of intima, and in places also of adventitia, concentric in arrangement; somewhere stenosed, many obliterated; liver was congested, showing some increase of connective tissue in yellowish areas.	Primary chronic endophlebitis of hepatic veins, possibly syphilitic, although nothing in the history or physical data suggesting syphilis.
Kelynack, 1897	32 years ♂	For two years abdominal pain, later ascites, œdema of feet, icterus; four last pregnancies ending in abortion.	Only one ostium of hepatic veins into vena cava present, and that admitted only a small probe and had thickened edges; firm adhesions of liver posteriorly at site of vena cava and also to diaphragm; nutmeg liver, enlarged, tough; spleen large; ascites; left hydrosalpinx.	Hepatic veins contained thrombi, some recent, others partly organized; portal-vein branches were "full;" liver cells degenerated, and in part necrotic; also an increase of connective tissue.	The stenosis of the hepatic veins appears to have been caused by an inflammation of the capsule and of the tissue at posterior surface of liver; the veins becoming stenosed by the dragging of the adhesions: possibly the process was of syphilitic origin.
Chiari (1) v. Maschka, 1899	28 years ♂	Previously always well; twelve days previous to death abdominal pain, and for the last fourteen hours continuous vomiting so that poisoning was suspected; delivered of healthy child five months previous to death.	The hepatic veins from their ostia to the smallest branches were filled with red and gray thrombi; their walls in the neighborhood of the vena cava showed circumscribed thickening; they admitted only the finest probe; liver was very large, surface smooth; spleen large; ascites; scar on under surface of labia major.	The walls of the hepatic veins were thickened throughout, the proximal parts showing marked inflammatory changes and a proliferating endo-and periphlebitis, extending a short distance along the vena cava; near the ostia, the endophlebitis caused stenosis by projecting elevations of newly formed tissue; liver congested, no increase of connective tissue.	Primary chronic inflammation of the proximal ends of the hepatic veins, probably syphilitic in origin.

Case.	Age and sex.	Clinical data.	Autopsy.	Microscopic data.	Conclusions of the author.
Chiari (2), 1899	55 years ♂	Woman had six children; five died in early youth; ill four weeks; ascites; œdema of legs, enlarged liver; sudden collapse.	The main trunks of hepatic veins entirely closed off by scar tissue from the vena cava, merely very small ostia being present; they contained thrombi, as did branches of portal vein; liver enlarged, capsule not thickened; spleen enlarged; hemorrhagic ascites and hemorrhagic infiltration of large intestine; partial aneurysm of left ventricle, obliteration of left coronary artery.	The ostia of the main trunks of the hepatic veins showed a proliferating endophlebitis: the vena cava showed near the openings slight thickening of the intima and in places adherent thrombi; the portal veins contained thrombi; the liver showed congestion and slight increase of connective tissue.	Primary obliterating inflammation of hepatic veins of long standing, the recent thrombosis causing the acute onset of symptoms, probably syphilitic in origin, for which the changes in the heart and coronary artery speak.
Chiari (3), 1899	29 years ♀	Abdominal pain, ascites, enlarged liver and spleen; death two months after onset; history of gonorrhœa.	Hepatic veins stenosed by thickening of their walls so that merely a probe could be inserted; the obstruction was increased by thrombi at the ostia; the liver enlarged, capsule somewhat thickened, cut surface showing a larger dark red area corresponding to diseased veins, and a smaller yellowish-brown area wherein the veins were merely slightly dilated; the branches of portal vein were dilated, but not thrombosed; spleen showed chronic enlargement; ascites; hyperæmia of stomach and intestines; hydrothorax.	The hepatic veins in their proximal and peripheral parts showed proliferating endophlebitis with thrombus formation, causing stenosis or complete obliteration; the portal veins were slightly dilated; the liver tissue in the dark area showed atrophy due to stasis, with slight increase of connective tissue and extravasation of blood; in the lighter areas, stasis was not present.	Primary endophlebitis of hepatic veins in the proximal and peripheral parts; secondary hepatic thrombosis with symptoms of stasis; possibly syphilitic in origin.
Churton, 1899	26 years ♀	Two years' history of enlargement of abdomen, and later distention of the superficial abdominal veins: not seriously ill until week before death; abdominal paracentesis showed blood-tinged fluid; history of trauma on right side previous to enlargement of abdomen.	The hepatic veins contained thrombi; one vein stenosed at entrance to vena cava contained an adherent thrombus, so that the ostium was difficult to find.	Hepatic veins obstructed by thrombi united to vessel wall by connective tissue; cirrhosis of liver with formation of "new bile-ducts."	Inclines to congenital syphilis as cause of stenosis of the hepatic veins and of cirrhosis of liver because no etiological factor could be found, and on the strength of successful mercurial treatment of a similar case.
Lazarus-Barlow, 1899	18 years ♀	Four months previous to exitus noticed swelling of abdomen, later melæna and hæmatemesis; on examination, superficial abdominal veins found distended, liver enlarged,	An hepatic vein was occluded by a mass of cicatricial tissue on the upper surface of liver posteriorly; the ostia were normal; the tissue extended 1½ inches into liver parenchyma, trabeculæ running from it into liver: the mass suggested a former gumma; numerous	Fibrosis of liver.	The hepatic veins obstructed by a cicatricial mass probably syphilitic in character; the thrombosis of the veins was secondary to the obstruction.

		Icterus present; tapped twice, sinking after second paracentesis.	branches of hepatic vein occluded by partially adherent thrombi; a localized suppurative peritonitis was present.		
Thran, 1899	32 years ♂	Two weeks previous to illness, gave birth to a child which died when three weeks old; complained of abdominal pain and enlargement; laparotomy for ovarian tumor revealed ascites and enlarged liver; later œdema of feet; four paracenteses performed.	Hepatic veins containing adherent thrombi; nothing said as to state of ostia; adherent thrombus 1½ cm. wide in inferior vena cava at diaphragmatic opening; liver small with greatly thickened capsule which sent broad connective-tissue bands into parenchyma; spleen much enlarged with thickened capsule; chronic parietal peritonitis.	Hepatic veins thickened, to a marked degree in parts, especially in inferior of liver; they also showed inflammatory infiltration, and almost all contained thrombi; from these veins connective-tissue strands could be traced to the liver capsule; these, however, not following the veins, but being irregularly distributed; liver extensively destroyed by growth of fibrous bands; portal veins not thrombosed.	The obstruction of the hepatic veins due to the pressure of the fibrous bands which resulted from the perihepatitis; the perihepatitis is thought secondary to a peritonitis probably gonorrhœal in origin.
Kretz, 1900	48 years ♂	Ascites for half-year previous to death; exploratory laparotomy revealed adhesions about gall-bladder and thickening of parietal peritoneum in bands; death three months later from marasmus.	Ostia of hepatic veins stenosed or obliterated by white endothelial masses, only four remaining intact; portal vein not affected; left lobe of liver small; right lobe large; the Spigelian lobe, projected almost like a tumor, was congested, and contained the normal hepatic veins running to vena cava.	Microscopic picture said to correspond to first of Chiari's published cases.	Thinks stenosis and obliteration of hepatic veins is due to exuberant scar tissue at the ostia of the hepatic veins, formed as result of trauma; frequently result of coughing; a second cause is diminished coagulability of blood in the hepatic veins; a third, the possible presence of infl'matory elements in the blood, as in syphilis or in infectious diseases.
Lichtenstern, 1900	20 years ♀	Since youth frequent attacks of diarrhœa and abdominal pain; a year previous œdema of feet; admitted twenty-three days previous to exitus, showing slight icterus, ascites, pain in epigastrium and loins; later œdema of feet and trunk, bloody urine passed, and toward end slight hæmoptysis.	The ostia of the two main hepatic veins and smaller upper veins obliterated, small ostia seen below, including those of veins going to Spigelian lobe; the obstructed veins contained adherent thrombi; portal veins normal; vena cava showed areas of thickening, and contained soft parietal thrombi; liver of normal size, capsule normal, contained red and yellow areas, the latter especially in region of portal trunk and in Spigelian lobe; ascites; spleen normal; hemorrhagic erosions of stomach; rectal hemorrhoids.	Hepatic veins obstructed by connective tissue at ostia; peripherally endo- and periphlebitis and organized thrombi; portal veins normal; vena cava showed signs of chronic inflammation; liver in red areas showed atrophy due to congestion and increase of connective tissue; in yellow areas cells were well preserved.	Hepatic veins closed by a primary inflammation of their walls leading to a seconday thrombosis; congenital syphilis probably the cause, although no other lesion of syphilis found.
Rendu et Poulin, 1901	35 years ♂	Abdominal pain and distention, ascites, dilated abdominal veins; death 3½ months after first symptoms; diagnosticat'd as cirrhosis of liver with thrombosis of vena cava.	Nothing said as to state of ostia of hepatic veins; vena cava and portal veins normal; liver somewhat enlarged, slight perihepatitis, marked cyanosis; great development of Spigelian lobe; spleen enlarged and indurated; thickening of peritoneum.	Marked congestion of liver with atrophy about the hepatic veins; hepatic veins showed obliterating endophlebitis and thrombosis.	Believes the phlebitis of hepatic veins due to alcoholism, but not sure whether it was primary or secondary to the thrombosis.

Case.	Age and sex.	Clinical data.	Autopsy.	Microscopic data.	Conclusions of the author.
Penkert, 1902	22 mos. ♀	Ascites, caput medusæ, enlarged liver and spleen; marked constipation.	Hepatic veins obliterated at ostia, slight retractions of wall of vena cava being present; the veins peripherally dilated and thrombosed; portal vein narrow, otherwise normal; liver enlarged, congested; capsule normal; anastomotic venous circulation.	Walls of hepatic vein thickened; liver congested, connective tissue increased, new formation and widening of biliary canals.	Obliteration of hepatic veins probably congenital; nothing pointing to syphilis.
Moore, 1902	25 years ♂	Ailing for three years; continuous pain in hepatic area for one month prior to admission; abdominal swelling noticed three days previous, also œdema of feet; dilated lower abdominal veins present; liver not palpable; no jaundice; eruption on legs suspicious of syphilis; paracentesis, followed by death five days later; ill five weeks in all.	Right and left hepatic veins only marked by shallow dimples and absolutely impervious for about one inch from ostia; they contained thrombi; liver small; no perihepatitis; it showed advanced central red atrophy; spleen small, congested; ascites; ductus venosus closed.	Hepatic veins closed by sparsely cellular connective tissue; media distinct, but showing intercellular fibrosis; intima thickened and obliterating the lumen; adventitia normal; hepatic veins thrombosed; liver showed central red atrophy, congestion, and hemorrhage.	Primary cause of obliteration of veins was endophlebitis, possibly due to syphilis; its localization to this area possibly due to the susceptibility of the veins in the vicinity of the ductus venosus to further obliteration.
Fabris, 1901	16 years ♀	Hemorrhagic ascites two months before death.	Large ostia of hepatic veins closed, only smallest existing; recent and older thrombi in hepatic veins; the vena cava somewhat whiter at site of obliterated openings; liver small, no perihepatitis; consistency increased; cut surface showed red zones and yellow zones. Caput medusæ.	No signs of proliferating endophlebitis of hepatic veins or of vena cava; walls normal; liver congested, with increase of bile-ducts.	Believes obliteration to be of congenital origin, as no reason to presuppose syphilis, and impossible to imagine an inflammation not leaving more evidence.
Meystre, 1901	28 years ♂	Three weeks previous to admission to the hospital complained of abdominal swelling; ascites found; operation, from which patient died two and a half months after onset of symptoms.	Hepatic veins contained thrombi; some were changed into connective-tissue bands, some showed thickened walls; the vena cava showed adherent thrombi overlying the obliterated ostia of the hepatic veins; some hepatic veins were patent; portal vein normal; liver congested and large; spleen enlarged; ascites.	Hepatic veins contained thrombi; some showed a thickened wall, the lining being irregular; in others the lumen was replaced by connective tissue; the vena cava inferior showed thickening of the intima at site of ostia of hepatic veins; portal veins normal; liver markedly congested, showed atrophy of parenchyma, combined with hypertrophy of some liver cells.	Primary endophlebitis of hepatic veins; no etiological factor present; author however inclines to syphilis as the cause.

An examination of the above columns shows that males and females have been affected in about equal numbers. Time of life brings with it no marked predisposition, as the ages of the patients have varied from seventeen months to fifty-five years, including the intervening decades; however, only three patients were over forty years of age. When we consider the clinical data, we find a general conformity in the course of the disease. The onset is generally gradual. Abdominal pain or discomfort, often most marked in the hepatic region, or an increase in the size of the abdomen, is almost invariably the first symptom, although for weeks or even months it may not be severe enough to cause the patient to seek medical advice. In some instances gastric symptoms have occurred later, but in a large majority these were not recorded. On examination the enlargement of the abdomen was found to be due to ascites, which rapidly increased and necessitated repeated paracenteses at short intervals. The fluid withdrawn had the characteristics of a transudate, but in 2 cases was slightly hemorrhagic. A caput medusæ, or distended abdominal vein, was often present. Œdema of the feet and anasarca occurred as late manifestations. Icterus was rarely noted, and then only of slight grade. The liver was invariably found enlarged, firm, and smooth in the early stage, but later small and slightly nodular. The spleen was large and firm. Temperature normal. Urine negative. The course of the disease rarely extended beyond six months from the date of onset. Such is the general clinical picture drawn from the 23 cases reviewed. The most marked variation from this type was the case of Lichtenstern, characterized by sudden onset and by gastric symptoms, resembling acute poisoning, followed by death within twelve days. When we review the conclusions, we find the greatest differences of opinion among the various authors. Most of them are in doubt, but each suggests a probable cause. Here, again, I believe, a table will most clearly show what these different opinions are:

	Perihepatitis.	Hepatitis.	Thrombosis.	Primary phlebitis.	Trauma.	Congenital anomaly.
Frerichs	2					
Rosenblatt	...	1				
Gee	...	1				
Schüppel	...	...	1			
Hainski	1					
Lange	...	...	...	1		
Kelynack	1					
Chiari	...	...	...	3		
Churton	...	...	...	1		
Lazarus-Barlow	...	1				
Thran	1					
Kretz	...	...	...	...	1	
Lichtenstern	...	...	...	1		
Rendu and Poulain	...	...	...	1		
Penkert	...	...	...	...	...	1
Moore	...	...	...	1		
Fabris	...	...	...	...	...	1
Meystre	...	...	...	1		
	5	3	1	9	1	2

A glance at the above table will be sufficient to show how varied are the views as to the cause of the disease. I shall not discuss the question of etiology at this point, but shall proceed to the consideration of the case which came under my personal observation.

The patient was a girl, aged sixteen years, in the general hospital at Prague, on the service of Professor Pribram, to whom I am indebted for the careful history. Family history negative; no indications of syphilis. Had always been pale and weakly, and had never menstruated. Had had none of the diseases of childhood. Four years previously she had been a patient in another division of the hospital, suffering from marked abdominal distention, causing dyspnœa. She remained there for six months, during which time the ascites gradually disappeared under diuretic treatment and after frequent paracenteses. She was discharged, with the diagnosis of hypertrophic cirrhosis of the liver. From that time until October 1, 1904, had performed light housework at home. She then began to suffer abdominal pain, without apparent cause, and within three days experienced marked swelling of the abdomen and distention of the veins of the upper abdominal wall. Although she left perfectly well otherwise, these symptoms induced her to apply for admission to the hospital. The results of the physical examination may be summarized as follows:

Face pale, lips and extremities cyanotic. Clubbing of toes and fingers. The veins of the thoracic wall appeared like a meshwork, from the midst of which a markedly distended vein extended toward the abdomen. The heart showed great pulsation and dilatation to the right; sounds muffled, pulse soft and rhythmic. Lungs negative, abdomen distended, the maximum circumference being 87 cm. The vein extending upon the abdomen from the thorax contained tender nodules. Movable fluid, extending to within 2 cm. of the umbilicus, could be clearly distinguished. Liver was palpable 2 cm. below free border of ribs, consistency firm, surface somewhat nodular. Spleen extended 1 cm. below free border of ribs. Inguinal glands not enlarged. External genitals infantile. Lower extremities not œdematous.

October 29th. Circumference of abdomen 89 cm. Diarrhœa.

31st. Abdominal pain. Sleeplessness. Diarrhœa. Afternoon temperature yesterday, 100.8°. Abdominal paracentesis with removal of 6 litres of slightly cloudy yellow fluid. The liver palpable three fingers below free border of ribs, hard, surface granular.

November 1st. Abdominal circumference 67 cm.

6th. Abdominal circumference 76 cm. Distention of veins includes upper abdomen, whereas yesterday it was confined to thorax.

7th. Abdominal circumference 78.8 cm.

8th. Abdominal circumference 79 cm. No œdema of feet.

9th. Severe abdominal pain.

13th. Death.

During her stay in the hospital the patient frequently had temperature as high as 100.8°. The urine was diminished in quantity and contained traces of albumin. The pulse tension (Gärtner) varied between 65 and 100. The treatment consisted in urea, theocin, and lecithin, at various periods, but all without effect.

Diagnosis. Cirrhosis hepatis and chronic peritonitis.

An autopsy was performed by Professor Chiari the day after exitus. The following is an abstract of the protocol: Body emaciated. Tonsils and cervical lymph nodes somewhat enlarged. Abdomen markedly distended; fluctuation present. The external genitalia were undeveloped and showed several superficial, firm, scars. From the mons veneris a bridge of skin, 3 cm. in length, stretched so as to form a furrow, extended to the clitoris. On opening the abdomen, greatly dilated subcutaneous and subperitoneal veins were apparent in the umbilical and in the epigastric areas. These ectatic veins were most numerous along the round ligament of the liver. Lungs adherent over entire surface; congested, slightly œdematous. Peribronchial nodes not enlarged. Pericardium contained 2 drachms of clear serum. Heart somewhat enlarged; the myocardium of light-brown color. Œsophagus dark red; its lower half showed markedly dilated submucous veins. The abdomen contained about 6 litres of clear serum. The peritoneum slightly thickened, smooth, glistening, especially thick over liver and spleen. Over the former it was "Zuckergussartig," or "iced," in patches. The omentum to a considerable extent adherent to the anterior abdominal wall in the epigastric area. It contained many dilated veins. Liver very small. Its width 20 cm., length 11 cm., thickness at point of greatest depth 7 cm.; its weight 950 gm. The surface showed numerous nodes, some 1 cm. in diameter. Consistency firm and tough. The cut surface of the liver showed everywhere numerous light-brown, circumscribed nodules, as large as a bean, projecting slightly from the surrounding parenchyma, which was grayish and of firmer consistency. No patent branches of the hepatic veins were apparent near the vena cava, but, instead, what seemed to be obliterated veins. The branches of the portal vein were patent throughout. The bile-ducts contained very dark bile. The trunk and main branches of the portal vein were dilated. The umbilical vein was present and patent for an ordinary anatomical sound. The intrahepatic portion of the vena cava was remarkable in that no ostia of the hepatic veins were visible on its inner surface. Ductus venosus arantii completely obliterated. Spleen weighed 170 gm., slightly enlarged. On section, tough, moderately vascular, trabeculæ soft. Malpighian corpuscles not evident to the naked eye. Kidneys moderately vascular, surface smooth, capsule non-adherent. In the perirenal tissue of the left side very numerous dilated veins, which could be traced into the mesentery of the descending colon. Mucosa of urinary bladder somewhat œdematous. Accessory ovaries present on both sides. Stomach contained stringy mucus; mucous membrane pale.

The middle third of the small intestine was intensely red, its wall greatly swollen by hemorrhagic infiltration, and nearly 1 cm. in thickness. This part as well as the adjacent lumen contained much fluid blood. The veins of the mesentery of this section were dilated and contained obstructing thrombi, which were reddish and friable. The arteries were normal. The splenic vein contained fluid blood. The mucosa of large intestine was slightly reddened. Pancreas and adrenals were pale.

The pathologico-anatomical diagnosis was: Obliteration of the hepatic veins at the ostia in the inferior vena cava. Obstruction of the hepatic veins in the liver. Atrophy of the liver. Dilatation of the submucous veins in the lower part of the œsophagus, of the subperitoneal veins, of the subcutaneous veins at the umbilicus, with persistence of the umbilical vein. Adhesion of the omentum to the anterior abdominal wall in the epigastric region, and dilatation of its veins. Hydrops ascites. Thrombosis of the mesenteric veins with hemorrhagic infiltration of part of the small intestine. Accessory ovaries. Scars of external genitals. Abnormality of vulva.

Before detailing the result of the microscopic examination, I wish to describe more fully the macroscopic picture of the liver and the hepatic portion of the vena cava. The conformation of the liver and the appearance of its cut surfaces were striking. Two points were noticeable at a glance: the relative smallness of the left lobe and the relative greatness of the Spigelian lobe, which, viewed from the posterior surface of the liver, projected downward like a tumor mass from the body of the organ. This abnormal development of the Spigelian lobe must not be looked upon as an accidental condition, as several of the reported cases showed a similar local development. It is to be considered, in a measure, characteristic. Closer examination of the various cut surfaces of the liver showed that the light-brown or yellowish nodules mentioned in the protocol were distributed throughout the organ and that they varied in size from a pinhead to 2.5 cm.; that they were not more numerous in the large right than in the small left lobe; that they were round or oval in shape, and were almost invariably surrounded by a definite ring of firmer tissue, macroscopically suggesting connective tissue. We next followed and examined the hepatic veins, the inferior vena cava, and the branches of the portal veins as they appeared successively in the various horizontally cut segments of the liver. The obliterated hepatic veins could without difficulty be traced into the parenchyma, in some segments for 2 to 3 cm., extending from the vena cava as firm, gray bands, without lumen. The vena cava in the posterior surface of the liver showed four minute ostia, ranging from a pinpoint to a pinhead in size. Three were at the upper angles of the vena cava, one to the left, two to the right, at the sites of the two main hepatic veins; the fourth was situated to the right of the median line at about the centre. The vena cava itself showed no stenosis. The portal vein likewise showed changes to the naked eye. Its right branch was obstructed by firm-contracting tissue not far from the porta hepatis; its left branch contained fresh thrombi, surrounded by reddish liver parenchyma.

The entire hepatic portion of the vena cava with the underlying liver tissue was removed for microscopic examination. Seven segments were thus obtained. Portions of the interior of the liver as well as of the spleen were also excised. All were hardened in alcohol and embedded in celloidin. Section after section was cut in order to follow an hepatic vein to its outlet, and to show the obliterated vessel in continuity with the vena cava. The Van Gieson and the hæmatoxylin and eosin stains were employed. However, a rapid examination of the unstained specimen often sufficed to show that the ostium of the vein had not been encountered. The obliteration was in all cases caused by connective tissue at the ostia, which was continuous with that of the intima of the vena cava. Up to this point the veins were to some extent patent. The obliterating connective tissue was dense, slightly cellular, and in parts contained lacunæ filled with blood. Careful search could not reveal any pigment therein, such as one should expect to find if the obliterating connective tissue had replaced a primary thrombus. The stenosis which existed in all cases beyond the point of obliteration was due to a proliferation of the intima. Here a thrombus was frequently found blocking the lumen. The media of the veins was well preserved, and ran in an uninterrupted course with that of the vena cava. Nowhere was a break therein to be seen as if trauma had caused a previous rupture of the fibres. The adventitia was also normal. The vena cava often showed a marked depression at the site of some obstructed ostia. Near these sites the connective tissue of its intima was thickened. At one point a partly organized thrombus was attached, but beneath it the connective tissue of the intima continued unbroken.

The liver parenchyma showed dilatation of the central veins, with atrophy of the liver cells in the centre of the lobules. There was a marked increase of connective tissue, which ran in bands through the parenchyma. Bile-ducts were numerous. The yellowish nodules, so striking in the macroscopic picture, proved to be composed of cells in a good state of preservation, showing no atrophy. They stained less deeply with eosin; but the nuclei took the hæmatoxylin well and were of average size. These areas were distinctive in showing no dilatation of the capillaries, a small rather than a large number of bile-ducts, and no increase of connective tissue. The spleen showed moderate congestion, but was otherwise normal.

From a clinical and a pathological standpoint this case is fairly typical. It must be regarded as one of long standing, having lasted at least four years. I consider that the patient was already suffering from an hepatic endophlebitis when in the hospital for the first time, although her disease was at the time diagnosed as hypertrophic cirrhosis. Later she improved, and the ascites remained away for the long period of four years, probably as the result of the compensatory circulation, which, as the autopsy showed, had established itself. The endophlebitis, however, continued, causing the stenosis of the ostia to become more and more marked, until finally obliteration occurred. This absolute closure of the hepatic veins, and barrier to the outflow of the large volume of venous blood from the liver, was the beginning of the end, and led to the sudden and persistent ascites which drained the strength of the patient.

The pathological findings serve to justify this interpretation. That we have to do with a case of long standing is borne out by the cirrhotic-like, granular liver, by the dense connective tissue obliterating its veins, and by the yellowish areas scattered throughout the parenchyma, which I look upon as regenerative areas, or so-called liver adenomata.

What cause can be assigned for this obliteration of the venous circulation, so strangely localized in one small area? No hereditary constitutional taint was disclosed; no previous illness of the patient existed upon which to cast the blame. Even the autopsy revealed little besides the venous obliteration and its immediate consequences upon the liver and the portal system. I believe that an obstruction of the hepatic veins can most certainly be caused by a perihepatitis, as happened in the cases reported by Frerichs[3] and others. But no such condition was found to explain this case. A primary thrombosis of the ostia I could not assume, partly because there was no reason for the formation of multiple thrombi in healthy veins, but still more because, as I mentioned above, no vestiges of a clot, no pigment, could be found at the site of obliteration. I have no doubt, however, that thrombosis also may be the primary cause of an obliteration through the formation of connective tissue. It has been urged by Kretz[20] that all these cases are due to trauma, to tearing of the hepatic veins, which are to be considered as constituting the insertion of the liver to the inferior vena cava; that small tears are brought about by coughing, and result in an exuberant scarring of the vessel wall, diminished coagulability of the blood in the hepatic veins, preventing thrombus formation. An argument of this nature cannot be discussed theoretically; everyone is entitled to his individual opinion as to the probable occurrence of such minute traumatic lesions. However, Kretz also bases his opinion upon the fact that microscopically he has seen a deflection and irregularity of the media of the obliterated veins at their insertion into the vena cava. With this point in view I examined the muscular coat of the veins most carefully, but could nowhere find a break in the media. In every instance I could follow it unbroken and undeflected into that of the vena cava. For this reason I cannot accept a traumatic origin for my case and must look for another cause. The etiological table given above shows that two cases have been ascribed to a congenital closure of the hepatic veins. To grant this as the etiological moment we should have to conclude that such a lesion might exist without symptoms for twelve years, or, if we apply it to many of the other published cases, well on into middle life. But what I believe furnishes absolute evidence against this origin is that the ductus venosus arantii was not patent. As Lazarus-Barlow[12] well says, this would not become obliterated under such abnormal conditions. Moore[19] and Rolleston[23] modify the congenital point of view by suggesting "a spread of the process of obliteration of the ductus venosus to the hepatic vein orifices—an excess of the obliterating process." This might well be true for the obliteration of the ostium of the left main hepatic vein, or for the vena cava at this point, but a study of the anatomical relations of this region does not allow me to believe that all the large hepatic ostia could be obliterated without causing some stenosis of the vena cava itself.

All the cases which I have considered above, as well as the one just reported have been uncomplicated by any narrowing of the hepatic vena cava. I now come to the last cause to be considered: a chronic endophlebitis. Clinically, my case and the majority of the cases, were slowly progressive in character. Pathologically, such change was found as is generally ascribed to chronic inflammation; a proliferation of dense connective tissue such as blocked the obliterated veins. Of all the causes advanced this one seems the most probable. We cannot, however, pronounce this view absolutely; as we are unable to define the exciting inflammatory cause. All but one of the writers who believe an inflammation to be the basis of the disease ascribe the changes to syphilis. However, to whichever opinion we incline, we must not lose sight of the fact that a close study of the cases shows that, with the exception of Chiari's first and, possibly, second case, none showed any other definite syphilitic lesions. If syphilis be regarded as one cause of the inflammation, it is probable that it is not the only one, but that inflammatory toxins of other natures can produce a like effect upon the lining of the hepatic veins.

As regards the diagnosis, I believe that I can venture the statement that no case of this disease has been correctly diagnosed. The cases have generally been confused with cirrhosis of the liver, although the ascites has also led to the diagnosis of tuberculosis or carcinoma of the peritoneum, and even to that of ovarian cyst. The two fulminating cases, characterized by vomiting, prostration, and a rapid course, were diagnosed as due to poisoning in the one instance and to intestinal obstruction in the other. It would lead to nothing to consider these cases further, as, in the present state of our knowledge, we could not hope to suspect the underlying condition when presenting itself in this violent form. But a study of the other cases shows that although this disease cannot be absolutely diagnosed, it can and should be thought of when a certain group of symptoms presents itself.

We have the typical signs of cirrhosis of the liver: ascites with dilated abdominal veins, sometimes a caput medusæ, very slight or no icterus, a palpable liver in the early stages, and a palpable spleen. What can lead us to suspect an obliteration of the hepatic veins as the cause? In the first place the history of the patient; the absence of all etiological cause for a cirrhosis. This is important. If, in addition to this, the patient is a very young individual, as is often the case, our suspicion should be awakened to the possibility that we are not dealing with an ordinary cirrhosis, a disease of late middle life.

Furthermore, an important differential diagnostic point is the occurrence of pain over the hepatic area, or localized in the upper abdomen, which is one of the most constant and earliest symptoms of endophlebitis, but exceptional in cirrhosis. Again, as Quincke[14] says, the rapidity with which the swelling of the liver and ascites develop is of importance. The patient with chronic endophlebitis will generally state that the swelling of the abdomen has showed itself within a few weeks or even a few days. In striking contrast to this is the gradual onset of abdominal swelling and ascites in cirrhosis. Later in the course of the disease the rapidity with which the ascitic fluid collects, the frequent necessity of paracentesis, point to an essentially obstructive lesion. All these symptoms, however, although brought about by a disease of the hepatic veins, are directly dependent upon the secondary obstruction in the portal system and are typical of portal obstruction. We cannot distinguish between them, and it is only the lack of cause for a portal obstruction that should make us consider whether this condition is not the result of a primary hepatic obliteration.

Beyond this we cannot go in the differential diagnosis. The paucity of cases presented for review does not allow of greater generalization. I am certain, however, that if this disease is taken into consideration at the bedside, and looked for in the postmortem room, we shall before long find many more upon which to base an opinion. Again, I wish to lay stress on the importance of opening the hepatic portion of the vena cava, and examining the ostia of the hepatic veins in all cases, but especially in those where ascites has been present.

BIBLIOGRAPHY.

1. Budd. Disease of the Liver, 1857.

2. Cohn. Klinik der embolischen Gefässkrankheiten, Berlin, 1860.

3. Frerichs. Klinik der Leberkrankheiten, Braunschweig. 1861. Bd. ii.

4. Rosenblatt. Ueber einen Fall von abnormem Verlauf der Lebervenen, Inaug. Dissert., Würzburg, 1867.

5. Gee. Complete Obliteration of the Mouths of the Hepatic Veins, St. Bartholomew's Hospital Reports, 1871, vol. vii.

6. Schüppel. Die krankheiten der Venæ Hepaticæ, Handbuch der spec. Path. u. Therap., v. Ziemssen, 1880, Bd. viii.

7. Hainski. Ein Fall von Lebervenen obliteration, Inaug. Diss., Göttingen, 1884.

8. Lange. Ibid., Kiel, 1886.

9. Kelynack. Stenosis of Hepatic Vein with Intrahepatic Thrombosis, Medical Press and Circular, London, 1897, vol. cxv., No. 25.

10. Chiari. Ueber die selbständige Endophlebitis obliterans der Hauptsämme der Venæ Hepaticæ als Todesursache, Ziegler's Beitr., 1899, Bd. xxvi pp. 1–18.

11. Churton. A Case of Cirrhosis of Liver apparently due to Congenital Syphilis with Thrombosis of Hepatic Veins, Transactions of the Pathological Society of London, 1899, vol. l. pp. 145, 146.

12. Lazarus-Barlow. Ibid., p. 146.

13. Thran. Ueber einen Fall von Lebervenen Thrombosis, Inaug. Diss., Kiel, 1899.

14. Quincke. Diseases of the Liver, Pancreas, and Suprarenal Glands, Quincke and Hoppe Seyler in Nothnagel's Encyc., Amer. ed., 1903, p. 903.

15. Lichtenstern. Ueber einen neuen Fall von selbständiger Endophlebitis obliterans der Haupstämme der Venæ Hepaticæ, Prager med. Woch., 1900, pp. 325–327, 338–339.

16. Kretz. Neisser's Stereoskopisch-medicinischer Atlas, No. 36, Leipzig, 1900.

17. Rendu et Poulain. Phlébite oblitérante d'une grosse Veine sus-hepatique; Apoplexie Hepatique, Bulletins et mémoirs de la Société médicale des hôpitaux de Paris, 6 Juin, 1901.

18. Penkert. Ueber idiopathische Stauungsleber (Verschluss der Venæ Hepaticæ), Virchow's Archiv, 1902, Bd. clxix. pp. 337–359.

19. Moore. Primary Obliterative Inflammation of the Main Trunk of the Hepatic Veins, Medical Chronicle, Manchester, July, 1902.

20. Kretz. Pathologie der Leber, Lubarsch-Ostertag Ergebnisse Allgem. Path. Abtheilung 2, Jahrgang 8, p. 498, 1904.

21. Fabris. Sulla Occlusione e trombosi delle vene epatiche, Archiv per le Sc. Med., 1904, Bd. xxviii. p. 221.

22. Meystre. Un Cas de Thrombose des Veines Hepatiques, Trav. de l'Institut de Stilling, Lausanne, 1904.

23. Rolleston. Diseases of the Liver, Saunders & Co., 1905.

A CASE OF EXCISION OF THE HEAD OF THE HUMERUS FOR CONGENITAL SUBACROMIAL DISLOCATION OF THE HUMERUS.[1]

By John B. Roberts, M.D.,
OF PHILADELPHIA.

A boy, aged three years, was operated upon on December 5, 1903. The head of the bone could be seen and felt under the acromial process, and the humerus was firmly abducted from the chest with the external condyle pointing almost directly forward instead of

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[1] Read by title at the meeting of the American Surgical Association, July 5, 1905.